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# Recombinant Human PCBD1 Protein (His Tag)

Catalog Number: PKSH032981

Note: Centrifuge before opening to ensure complete recovery of vial contents.

### Description

Species Human

Source E.coli-derived Human PCBD1 protein Ala2-Thr104, with an N-terminal His

Calculated MW14.2 kDaObserved MW14 kDaAccessionP61457

**Bio-activity** Not validated for activity

#### **Properties**

**Purity** > 95 % as determined by reducing SDS-PAGE.

**Concentration** Subject to label value.

Endotoxin < 1.0 EU per µg of the protein as determined by the LAL method.

**Storage** Store at < -20°C, stable for 6 months. Please minimize freeze-thaw cycles.

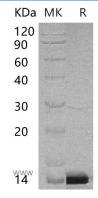
**Shipping** This product is provided as liquid. It is shipped at frozen temperature with blue ice/gel

packs. Upon receipt, store it immediately at < - 20°C.

Formulation Supplied as a 0.2 μm filtered solution of 20mM Tris-HCl, 150mM NaCl, 1mM DTT,

pH 8.0.

#### Data



> 95 % as determined by reducing SDS-PAGE.

## Background

Pterin-4-α-Carbinolamine Dehydratase (PCBD1) is the founding member of the Pterin-4-α-Carbinolamine Dehydratase Family. PCBD1 is involved in Tetrahydrobiopterin biosynthesis. It seems to prevent the formation of 7-Pterins and accelerate the formation of Quinonoid-BH2. Furthermore, PCBD1 regulates the homodimerization of the transcription factor Hepatocyte Nuclear Factor 1 (HNF1) and enhances its transcriptional activity. Defects in PCBD1 are the cause of BH4-Deficient Hyperphenylalaninemia Type D (HPABH4D). HPABH4D is characterized by the excretion of 7-substituted Pterins in the urine of affected patients.

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